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Obstruction of Right Ventricular Inflow and Outflow in Corrected Transposition of the Great Arteries {S,L,L}: Two-Dimensional Echocardiographic Diagnosis

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Obstruction of systemic ventricular inflow and outflow is considered uncommon in corrected transposition of the great arteries {S,L,L}. Between 1979 and 1985, 42 patients with corrected transposition and two ventricles and atrioventricular valves underwent two-dimensional echocardiography. Obstruction of right ventricular inflow and outflow was present and diagnosed by two-dimensional echocardiography in 5 of the 42 patients. A supratricuspid stenosing ring, recognized in the apical or subxiphoid four chamber view as a bright, linear structure on the left atrial side of the tricuspid valve, occcurred in two patients. Subaortic obstruction due to infundibular hypertrophy with or without displaced muscle bundles was seen in three patients. Subxiphoid long- and short-axis views and parasternal long-axis views best displayed these features. Aortic coarctation was present in four cases and could be diagnosed using modified suprasternal notch views.

Thus, systemic ventricular inflow and outflow obstruction may be more common in corrected transposition than previously believed (occurring in up to 10 to 15% of patients). The mechanisms producing the obstruction appear to be characteristic of the left atrium and right ventricle irrespective of location or connections. Echocardiography appears to be an excellent technique for diagnosing these associated lesions in corrected transposition.

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Although pulmonary stenosis is common in corrected transposition of the great arteries {S,L,L}, obstruction of inflow and outflow of the systemic ventricle (anatomically the right ventricle) is uncommon. Nonetheless, recognition of this complicating feature may be crucial for successful surgical repair. Although isolated case reports (1–7) of obstruction on the systemic ventricular side in corrected transposition exist, the larger clinical (8–15), angiographic (16,17) and echocardiographic (18–21) series generally do not describe this feature. A single autopsy series (22) described both inlet and outlet obstruction of the systemic right ventricle in corrected transposition. We herein report the two-dimensional echocardiographic findings in five patients with corrected transposition and obstruction of systemic ventricular inflow or outflow.

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Methods

Subjects. Between 1979 and 1985, 42 patients with corrected transposition of the great arteries {S,L,L}, with two ventricles and atrioventricular (AV) valves, underwent two-dimensional echocardiographic examination at this institution. Five (12%) of these patients were found to have obstructive lesions of systemic ventricular inflow or outflow and are the subjects of this report (Table 1). Their ages ranged from 2 days to 23 months at the time of examination; four patients were male and one was female. The examination was performed before surgery in three patients and after pulmonary artery banding or coarctation repair, or both, in two other patients.

Echocardiography. Two-dimensional echocardiography was performed using a Picker Echoview 80 CI, a Diasonics Cardiovue 100, an ATL Mark 600 or a Hewlett-Packard 77020 Cardiac Imager equipped with a 5 MHz transducer. Subxiphoid, apical, parasternal and suprasternal notch views were obtained in each case. The examinations

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Table 1. Associated Malformations in Five Patients With Corrected Transposition and Systemic Ventricular Obstruction

		Inlet Obstruction	Outlet Obstruction				
Case	Age	Supratricuspid Stenosing Ring	Subaortic Stenosis	Aortic Coarctation	Ebstein's Anomaly	VSD	Subpulmonary Stenosis
1	23 mo	+	−	+	−	−	+
2	13 mo	+	−	−	−	+	+
3	4 days	−	+	+	+	−	−
4	5 mo	−	+	+	−	+	−
5	2 days	−	+	+	+	+	−

VSD = ventricular septal defect; + = present; − = absent.

were recorded on 1/2 inch (1.27 cm) video cassette tape for review in real time, slow motion and stop frame modes. Cardiac anatomy was described using nomenclature proposed by Van Praagh et al. (23).

Results

Details of cardiac anatomy in these five patients are shown in Table 1.

Right ventricular inflow obstruction. A supratricuspid stenosing ring (22,24–26) was identified prospectively in two patients (Cases 1 and 2) as a bright linear echo just above the left-sided tricuspid valve (Fig. 1A and B). In both cases this fibrous ring inserted medially onto the left atrial surface near the crux of the heart and laterally onto the left atrial free wall between the left atrial appendage and the tricuspid valve anulus. Doppler examination in both cases disclosed mild tricuspid stenosis (peak pressure drop of 8 to 10 mm Hg) which was confirmed by direct pressure measurement at cardiac catheterization.

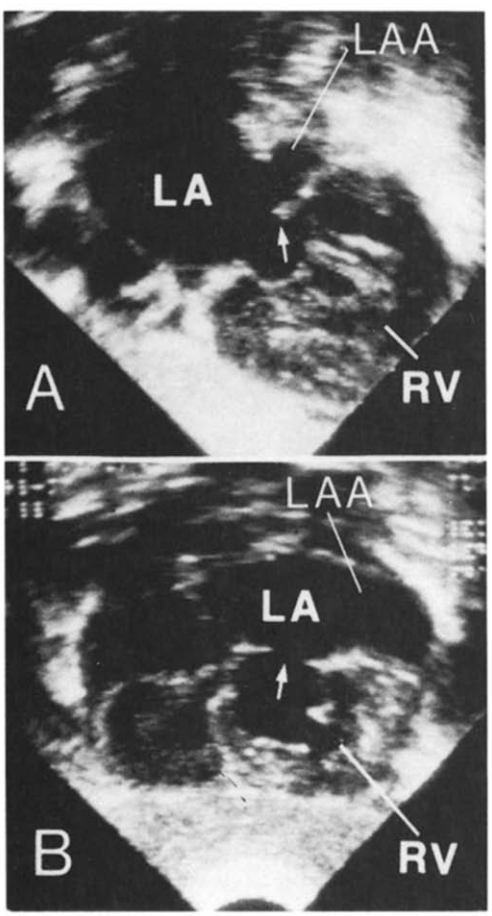
Right ventricular outflow obstruction. *Subaortic stenosis was diagnosed prospectively in three infants (Cases 3 to 5)* using parasternal and subxiphoid long-axis views (27,28). In two cases with a ventricular septal defect (Cases 4 and 5) the obstruction was due to anterior and leftward displacement of the infundibular septum in addition to hypertrophy of the infundibular free wall (Fig. 2). This combination produced a situation similar to that found in double chamber right ventricle (29). In Patient 4 a subaortic pressure gradient of 20 to 30 mm Hg was confirmed by Doppler examination and cardiac catheterization.

In the patient with an intact interventricular septum (Fig. 3A), marked hypertrophy of the infundibulum without cavity expansion produced extreme right ventricular outflow tract obstruction. The ascending aorta in this case was severely hypoplastic, as seen in hypoplastic left heart syndrome.

Aortic coarctation was present in four patients (Cases 1 and 3 to 5). The diagnosis was made prospectively by two-dimensional echocardiography in two patients (Cases 3 and 5) using a suprasternal notch or high left parasternal view (30,31). Because of the levoposition of the ascending aorta in corrected transposition, the transducer must be rotated counterclockwise about 30 to 45° from the usual suprasternal notch long-axis view to display the aortic arch (32) (Fig. 3B).

The other two patients (Cases 1 and 4) were first examined after repair of coarctation. In one patient severe residual coarctation was correctly diagnosed and in the other

Figure 1. Subxiphoid four chamber view in Cases 1 (**A**) and 2 (**B**) showing the supratricuspid stenosing ring (**arrow**). LA = left atrium; LAA = left atrial appendage; RV = right ventricle.



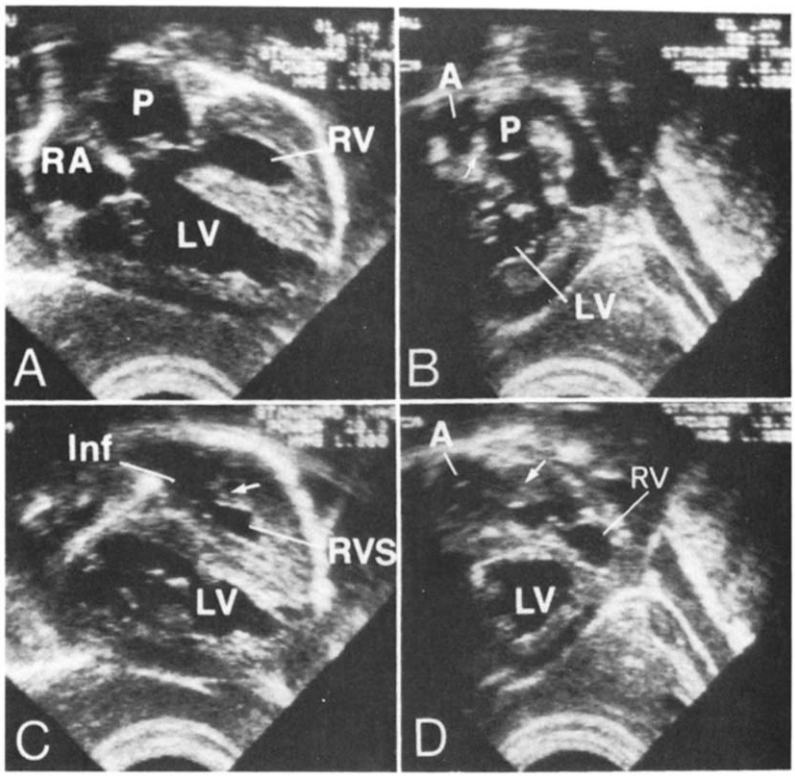


Figure 2. Subxiphoid views in Case 4. **A**, Coronal plane view showing a large subpulmonary ventricular septum and overriding of the pulmonary artery (P). **B**, Parasagittal plane view showing the infundibular septum (**arrow**) displaced anterosuperiorly toward the right ventricle (RV). **C**, A more anterior coronal plane view showing the "muscle bundles" (**arrow**) dividing the inflow and outflow portion of the right ventricle. **D**, Parasagittal plane view with slight clockwise rotation illustrating the role of the infundibular septum (**arrow**) in producing the midchamber obstruction of the right ventricle. A = aorta; Inf = infundibulum; LV = left ventricle; RA = right atrium; RVS = right ventricular sinus.

the characteristic "kinking" of the aortic isthmus subsequent to coarctectomy could be seen.

Associated abnormalities. Ebstein's malformation of the left-sided tricuspid valve was demonstrated in two patients (Cases 3 and 5) using apical and subxiphoid views (18,21). Severe tricuspid regurgitation was present in Patient 3.

A large perimembranous ventricular septal defect was seen in Patient 2. As noted earlier, a conoventricular septal defect with anterior deviation of the infundibular septum and overriding of the interventricular septum by the pulmonary artery were present in Patients 4 and 5. The right ventricle was somewhat hypoplastic in two patients (Cases 3 and 5). Subpulmonary stenosis was present in both patients with a supratricuspid stenosing ring.

Discussion

The cardiac malformations most commonly associated with corrected transposition are ventricular septal defect, pulmonary stenosis and tricuspid valve anomalies (12–15). Obstruction of systemic (right) ventricular inflow or outflow in corrected transposition has been reported in only a few cases (1–7) and is not generally considered a component of this lesion. However, our findings as well as those of another recent report (22) demonstrate that systemic ventricular obstruction occurs in up to 10 to 15% of patients with corrected transposition.

Morphologic features of obstructive lesions. The morphologic features of the obstructive lesions seen in our patients deserve special attention. The supratricuspid stenosing

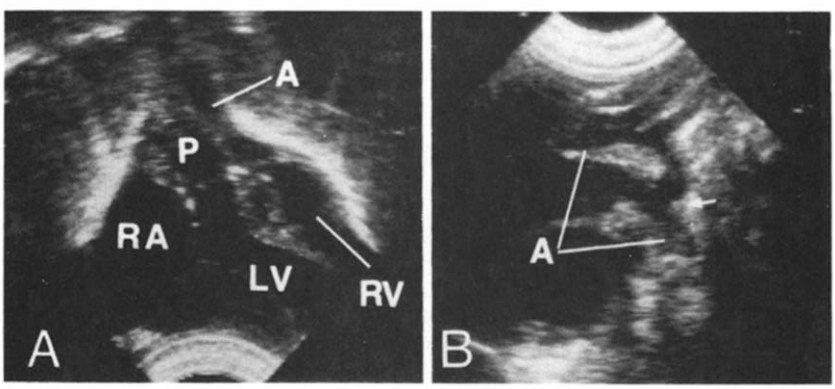


Figure 3. Subxiphoid and suprasternal notch views in Case 3 with Ebstein's anomaly of the tricuspid valve, intact ventricular septum, severe infundibular stenosis and hypoplastic aorta. **A**, Anterior coronal plane view showing the narrowed subaortic infundibulum and hypoplastic ascending aorta (A). **B**, Suprasternal notch view of aortic arch (A) showing the juxtaductal "shelf" (**arrow**) at the junction with the ductus arteriosus typical of coarctation. Abbreviations as in Figure 2.

ring, although uncommon in AV discordance, has been reported previously (22,24–26). This structure appears remarkably similar to the supramitral stenosing ring reported by Shone et al. (33) in patients with AV concordance and multiple left heart obstructive lesions. Recognition of this structure in the usual left atrial position despite AV discordance suggests that the supravalvular ring is a form of atrial outlet obstruction peculiar to the left atrium, irrespective of the morphology of the AV valve through which it empties.

The mechanisms of subaortic obstruction in our patients are similar to those seen in right ventricular outflow tract obstruction with a ventricular d-loop and normally related great arteries (28) or common transposition (34,36). Anterior displacement of the infundibular septum or hypertrophied infundibular muscle bundles, or both, as seen in tetralogy of Fallot, double chamber right ventricle or common transposition with subaortic stenosis, produced narrowing at the infundibular inlet. Generalized infundibular hypoplasia was also seen in one case.

Conclusions. Systemic ventricular inflow or outflow obstruction in corrected transposition is more common than was previously appreciated. The types of obstruction seen appear to be characteristic of the left atrium and right ventricle irrespective of location or connections. Two-dimensional echocardiography is an excellent technique for detecting these associated obstructive lesions.

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